BARTHOLOW (R.)

ON THE

PROGRESSIVE LOCOMOTOR ATAXIA:

ITS

HISTORY, SYMPTOMATOLOGY, PATHOLOGY, AND TREATMENT.

BY

ROBERTS BARTHOLOW, A. M., M. D.,

Professor of Physics and Medical Chemistry in the Medical College of Ohio; Lecturer on Clinical Medicine and Physician to St. John's Hospital; formerly Assistant Surgeon (Captain) U. S. Army, etc.

CINCINNATI:

ROBERT CLARKE & CO., 55 WEST FOURTH STREET.

1866.

Lancet & Observer Compliments of Dr Bartholow

ON THE

PROGRESSIVE LOCOMOTOR ATAXIA:

ITS

HISTORY, SYMPTOMATOLOGY, PATHOLOGY, AND TREATMENT.

BY

ROBERTS BARTHOLOW, A. M., M. D.,

Professor of Physics and Medical Chemistry in the Medical College of Ohio; Lecturer on Clinical Medicine and Physician to St. John's Hospital; formerly Assistant Surgeon (Captain) U. S. Army, etc.

CINCINNATI:

ROBERT CLARKE & CO., 55 WEST FOURTH STREET.

1866.

Neque enim credunt, posse eum scire, quomodo morbos curare conveniat, qui, unde ii sint, ignoret.

A. Corn. Celsi *Medicinæ Liber I.*

Non minus certo etiam a minutissimis morbi circumstantiis indicationes curativas possit Medicus desumere, quam ab iisdem sumpsit diagnostica.

Sydenham. *Ob. Medicæ Præf. Ed. Tert.*

PREFACE.

In the preparation of this little essay, I have made liberal use of the elaborate work, *De L'Ataxie Locomotrice, et en particulier de la maladie appellée Ataxie Locomotrice Progressive*, by Dr. Paul Topinard. The sections on the history and symptomatology have been condensed from that work chiefly. I have added some historical facts and references, and have contrasted the symptomatology of my patient with that described by Dr. Topinard. Whatever aid has been derived from other sources is duly acknowledged.

This essay was written for the *Cincinnati Journal of Medicine*, and appeared in the numbers for April, May, and June. I had the conviction, in preparing it, that the readers of that periodical would not be unthankful for some recent information on a rare form of nervous disease, which, as it happened to me, might also happen to them. I do not urge the novelty and importance of my labor in justification of its republication in this form. It is reprinted now for distribution among such of my personal friends and of the professional public as may be interested in studies of this character.

R. B.

344 Race Street, Cincinnati, O.

PROGRESSIVE LOCOMOTOR ATAXIA:

ITS

HISTORY, SYMPTOMATOLOGY, PATHOLOGY AND TREATMENT.

NOMENCLATURE.—The *Ataxie Locomotrice Progressive;* Duchenne's Disease; Progressive Spinal Paralysis; Tabes Dorsalis; Progressive Locomotor Ataxia, etc.

Stewart Kelch, by occupation a gilder, was admitted to St. John's Hospital, service of Prof. Bartholow, Feb. 10, 1866.

He gave the following history:

About the 15th of December last he began to experience some troubles of vision; objects were confused and indistinct; soon after he had double vision. He noticed that his food had no taste about the same time. His hearing and smell were unaffected. In consequence of these ocular troubles he consulted a female doctor who had some reputation as an oculist, who prescribed an eye-wash and some medicine. Finding that no improvement took place he consulted a well-known oculist in this city, who, after a prolonged ophthalmoscopic examination, directed glasses and gave him some internal remedy. His eyesight, soon after this, suddenly improved.

Whilst he was in attendance upon the female doctor, he began to experience numbness and lack of voluntary control in the inferior extremities, and the apparent loss of power and the difficulty of locomotion were well marked during the period he was under the care of the oculist, who, probably, did not recognize the nature of the malady.

Soon after troubles of the same character appeared in the upper extremities.

During the whole period he suffered from nocturnal emissions, satyriasis and at length complete impotence. He had had some troubles in micturition and defecation, the desire coming on suddenly and the evacuation occurring immediately.

He had no rheumatic pains. Pricking and tingling in the extremities and a dull pain in the loins and limbs, which appeared about the time his ocular troubles began, were the only symptoms of this kind.

Symptoms on admission.—Kelch is 36 years old, 5 *ft.* 6 *in.* high, light complexion, lymphatic temperament, full face with prominent lower jaw; very protuberant blue eyes.

He has no troubles of vision at present except some dimness of sight; his eyes are more prominent than natural. His face is expressionless. His taste has returned, but his tongue is protruded to the right. Sensibility, both tactile and muscular, diminished in the upper extremities, especially the right. Power of co-ordinating movements not completely lost, but diminished, in consequence of which his attempt to grasp any object is uncertain and vacillating. He can not use his knife and fork. He complains especially of a sensation of numbness in his hands and forearms, and a sense of pricking. They are, .5° F., colder than natural. There is no wasting of the muscles, but a contraction of the central tendon of the *extensor communis digitorum*. His grasp is as strong nearly as in health.

When requested to stand with his feet together and his eyes closed. he can not do so without the support of a chair. He walks as an unpractised youth on stilts. When requested to walk across the room, he rather runs than walks, and evidently can not control the movements of his feet, which are thrown up and down and laterally, in the most irregular manner. The chief sensation in his feet is that of numbness. Tactile sensibility and myotility are more diminished in lower than upper extremities. He perceives the galvanic shock when a strong current is passed through his limb, but feebly. The muscular sense is almost abolished. In walking, he can scarcely feel the floor, and when his eyes are not fixed upon his feet, he can not direct his movements.

He has been for some weeks entirely impotent.

His intellect appears to be unimpaired.

Treatment.—He was put upon the crystallized nitrate of silver, 1-6 of a grain *ter in die*. After ten days' use of this remedy, I gave him 30 grains of the iodide of potassium daily. His improvement under this treatment has been most marked. He is very rapidly regaining the power of co-ordination of muscular movements, his cutaneous and muscular sensibility are returning; he walks with considerable vigor

and certainty, and he begins to experience sexual desire with erections. The iodide of potassium seemed to be indicated by the fact that the patient had for ten years followed the profession of a gilder, and had during that period of time, absorbed more or less of the mineral matter used in the prosecution of his trade. As M. Melsens has demonstrated that this remedy makes soluble combinations with these mineral poisons, thereby facilitating their excretion, it was applicable to the treatment of this case on the theory that the alteration in the spinal cord might be due to the slow and long continued action of the minerals. Moreover, the iodide of potassium is one of the most reputable remedies in this affection.*

HISTORY. The name, *ataxie locomotrice progressive*, was given this malady by Duchenne of Boulogne, who described it with more particularity than any of his predecessors, but he is not entitled to the merit of discovery. It had been recognized in England in 1847, and was well described in Germany in 1834, but under other names, as paraplegia with disordered co-ordination of movement, *tabes dorsalis*, gray degeneration of the posterior columns of the spinal cord, progressive spinal paralysis, etc.

The most ancient of these terms, *tabes dorsalis*, originated with Hippocrates, who applied it to the accidents produced by the abuses of venery. The passage in which the term occurs is found in *De Morbis*, usually ascribed to Hippocrates, but by Dr. Adams, translator of the the Sydenham Society edition of the works of Hippocrates, is assigned to the Cnidian school: *Tabes dorsalis a spinali medulla oritur, maxime vero recentes sponsos et libidinosos corripit.*

Sauvages, a long time after, described a group of symptoms, produced by the same cause, which closely correspond to the disease now known as progressive locomotor ataxia. Lallemand also, under the title of the "dorsal consumption," described a set of symptoms of the same character.†

The term *tabes dorsalis* was, by the Germans, changed in its signification, and applied to a disease of the cord characterized by atrophy of this organ. The first autopsy, clearly establishing this condition of the cord, was made so long ago as 1679. The disease was very clearly described in Hufeland's Practice of Medicine, under the name tabes dorsalis, in 1834. Steinthal, in 1844, was equally explicit in his details concerning this disease, describing, as had Hufeland, the difficulties of

* See reports of two cases in the *Medical Times and Gazette*, treated with this remedy, Jan. 27, 1866. They were only improved, however.

† *Des pertes séminales involontaires. Paris et Montpelier*, 1839-41.

locomotion, the loss of power of co-ordinating muscular movements, the amaurotic amblyopia, etc.

Romberg,* in 1851, under the same name of tabes dorsalis, and Wunderlich, the following year, under the title of progressive spinal paralysis, particularize with great precision the essential phenomena of this disorder.

The morbid anatomy of progressive locomotor ataxia had been studied with the naked eye only, prior to 1857, in which year Ludwig Turck published a memoir at Vienna, in which he described the microscopic appearances of the degenerated posterior columns of the spinal cord. Virchow and Raciborski confirmed these observations, which had established that the alteration proper to *tabes*, consisted in an atrophy of the nervous elements with hypertrophy of the intermediate connective tissue.

Whilst these observations were accumulating in Germany as to the nature of tabes dorsalis, attention was being called in England to defects of co-ordination, dependent upon an affection of the cord. Todd, especially, influenced by the theoretical views which he entertained as to the functions of the posterior columns of the spinal cord, and having had two cases in which defects in the co-ordination of voluntary movements existed with integrity of the muscular force, diagnosticated during life and confirmed by *post mortem* observation, a lesion limited to the posterior columns. "Two sorts of paralysis of movement," says Todd, "are found in the inferior extremities; one consists in feebleness or loss of voluntary movement; the other is distinguished by diminution or complete abolition of the power of co-ordination of movements." He says nothing, however, of ocular troubles.

In 1856 and 1858, Gull published in Guy's Hospital Reports a long series of observations, relative to diseases of the cord. In one case he observed trouble in the co-ordination of movements and alterations of the posterior columns of the cord. Gull, ignoring the labors of the Germans in the same field, and the investigations of Duchenne not having yet seen the light, did not hesitate to ascribe these phenomena to chronic myelitis.

In France various observations were made and recorded in the proceedings of the anatomical society, and in the immortal work of M. Cruveilhier. These observations extend from: 1828, those of M. Hutin; 1830, those of M. Cruveilhier; 1845, those of M. Fredault; 1847, those of Monod; 1856, those of M. Luys; to 1858, those of M. Laborde.

---

* Manual of Nervous Diseases. *Syd. Soc. Trans.*

It is subsequent to these observations that M. Duchenne announced in the *Archives de Médicine* of 1858, the existence of a new disease which he proposed to call "*ataxie locomotrice progressive*," describing it in these terms: "Progressive abolition of co-ordination of movements and apparent paralysis, contrasting with integrity of the muscular force." This able observer ignored the ideas of Todd and the labors of Hufeland, Steinthal, Romberg and Ludwig Turck. His merit consists in this: that he comprehended the relation of the symptoms, their habitual succession, and insisted more than Romberg had done, upon the ocular troubles. This was certainly the view of the merit of Duchenne entertained by Trousseau, when he baptized the new disease by the name of its presumed discoverer.

SYMPTOMATOLOGY.—The duration of this malady is from one to thirty years; hence, it is useful to make some divisions to facilitate the description. We adopt the division of M. Duchenne: "The *ataxie locomotrice progressive* is divisible into three periods: the first characterized by three symptoms—pains, ocular troubles and anaphrodisia; the second, by disorders of muscular sensibility and of the sensibility of the inferior extremities; the third, by the extension of the same troubles to the superior extremities."

Pains.—These are in general the first symptoms to attract the attention of the patient.

In one hundred and four observations made with reference to the frequency of the pains and the period of their appearance relative to the ataxic symptoms, they were present in forty-four before the locomotor ataxia; in twenty, after; in eighteen, present, but time not definitely fixed; in twenty-two, absent.

They were present in the case of Kelch after the locomotor ataxia.

These pains are observed in all diseases of the cord, but are more frequent in progressive locomotor ataxia.

At their origin, these pains are fugitive and distributed over the surface of the body or one of its lateral halves, or they are limited to one region, to one extremity, especially to the feet. If, after having been generally distributed, they disappear in the superior half of the body to concentrate in one limb, this part, in which a primitive or secondary localization of the pains has taken place, is destined to be the first attacked by the ataxic disorders. They are sometimes superficial, sometimes deep-seated, in the muscles or in the neighborhood of the large joints. They are encountered in the head, jaws, orbital cavities, external auditory canal, the urethra, the deeper parts of the pelvis. In the globe of the eye they give rise to congestive phenomena, identical with

those that accompany neuralgia of the fifth; injection of the conjunctiva, lachrymation, heat, dilatation of the pupil.

These pains are resolvable into two classes: the one, dull and nearly continuous, exceptional; the other, short and sharp, habitual.

The first have been compared to the sensation produced by the constriction of a limb by a cuirass.

The second, are sharp pains in the extremities shooting toward the trunk; more rarely, they are likened to flashes of fire, puncture of an awl, strokes of a hammer, or vibrations flowing along in the direction of the muscles, or more commonly, as a *tic tac* in some point like that of a watch.

Their fundamental character is their mobility and intermittence. They may appear and disappear many times a day, and the attacks vary in duration from some hours to seven days or more. It is exceptional for them to increase at night. Sometimes the pains are so horrible as to justify the expression of Remak: *tabes dolorosa*.

Their analogy with certain rheumatismal pains is remarkable. They are influenced by all those atmospheric causes which exasperate rheumatic pains. Hence it is that winter is a bad season for the ataxic.

Functional troubles of the cranial nerves.—These functional derangements of the cranial nerves were well marked in our case. He had amblyopia, diplopia, protrusion of the eye, paralysis of taste, etc. The comparative frequency of these functional troubles of the cranial nerves—optic, motor oculi, hypoglossal, auditory, facial, glosso-pharyngeal, etc., without regard to period is as follows: present in ninety-seven cases; absent in twenty-eight.

The cranial nerves are divisible into two or three orders: the first preside over movement; the other over special sensibility, but some of their filaments have general sensibility. What is the nature of the disorder proper to each?

It is evident that the alterations of the optic, auditory, lingual, trigeminal (sensitive part), produce but two orders of phenomena: hyperæsthesia (the neuralgic or rheumatic pains already alluded to); special anæsthesia (amblyopia, paralysis of taste).

In regard to the motor oculi, facial, inferior maxillary, hypoglossal, that portion of the pharyngeal plexus which presides over the muscular sensibility of the vail of the palate, and of the pharynx, and the recurrents, their functions will be exalted, or abolished, or perverted; the muscles to which they are distributed will be either paralyzed or ataxic.

The phenomena proper to the motor oculi are constantly paralysis, except two cases in which Friedreich saw double bilateral nystagmus, and one in which M. Beau speaks of defects in the co-ordination of the movements of the eyes.

Facial hemiplegia, when there exists greater or less irregularity in the two sides of the face, has been noticed.

Embarrassment of speech and pronunciation has been observed in sixty-seven cases.

The time when these functional troubles of the cranial nerves appear varies. The ocular troubles in more than half of the cases appear in the first period, a little after the pains and sometimes in their absence; the feebleness of vision and the embarrassment of speech appertain to the second period; and the dysphagia and the paralysis of the vail of the palate to a still more advanced period. The most ephemeral and intermittent of the ocular troubles are the diplopia and strabismus. The temporary character and spontaneous disappearance of these accidents give rise to a therapeutic illusion. Thus, the subject is surprised by them in good health; the pains which precede or accompany them are referred to rheumatism; the connection between the two is not understood. He goes to an oculist who diagnoses congestion of the papilla or paralysis of the third pair, orders an unguent and collyria, electrizes the orbital cavities, and as if by enchantment the malady disappears. These examples are very common. It is the history of our own case. Did the oculist effect a cure? The symptoms would have disappeared as well with cold water.

The disorders pertaining to the orbital cavities are paralysis of motor oculi, producing ptosis, external strabismus, diplopia, and dilatation of the pupil; paralysis of the motor externus, producing internal stabismus and diplopia; paralysis of the patheticus, producing also diplopia and strabismus and other vague symptoms. Finally, kopiopia, amblyopia, and amaurosis occur. The diplopia is generally attributed to defect in the convergence of the two visual axes upon the object examined.

The condition of the retina, according to the ophthalmoscopic examinations, consists in congestion or atrophy, more or less advanced, of the optic papilla. This is confirmed by some post-mortem observations; in others, however, no alteration is found after death. The absence of lesions, remarked by various authors, in the cranial nerves affected with functional troubles, is in conformity with the temporary character of the different symptoms. It is this fugitiveness which

enables the distinction to be made between them and similar maladies arising from other causes.

Troubles in the genital organs.—In respect to importance, the disorders of the genital organs take place after the pains and ocular derangements. In Germany especially, where the ancient views on the subject of *tabes dorsalis* are not yet forgotten, a particular interest attaches to these symptoms. That venereal excesses are causative can hardly be admitted. As it is difficult to question females, all our observations are silent with regard to them. Moreover, nocturnal pollutions may occur without constituting a morbid state. Finally, the subjects who are attacked by this malady have generally attained the age of from thirty-five to forty years, the period of natural decline in the sexual desire.

Four symptoms present themselves: spermatorrhœa, anaphrodisial and impotence. The first occurs among the earliest symptoms of the first period. In the case of Kelch, spermatorrhœa had existed for six months prior to the occurrence of the ocular troubles. He admitted to venereal excesses. The nocturnal pollutions, accompanied at first by erection and pleasurable sensations, finally became passive. After the spermatorrhœa, or sometimes preceding it, there is a progressive diminution of the venereal desires, difficulty in satisfying them, and at length complete impotence.

Vesical disorders are observed in some cases. The trouble consists in slow emission of the urine, incompleteness in emptying the bladder, and inability to determine when the emission of urine has terminated. Incontinence has been observed, but retention rarely.

Among the disorders of micturition is found anæsthesia of the vesical and urethral mucous membrane, in consequence of which the individual is unconscious of the emission of urine. The dysuria, as the ocular paralysis, is temporary and intermittent.

Rectal troubles are rare. Constipation is habitual. When the matters are liquid they escape before the patient has time to reach the closet. Incontinence does not occur until about the commencement of the second period.

These vesical and rectal troubles occurred in our patient Kelch.

Duchenne's Second Period.—The boundary between the first and second period is not so clearly marked as the arbitrary division of Duchenne would indicate. It is by very gradual and sometimes almost imperceptible steps that the patient passes from one to the other, although but a few months may be occupied in the transition. It is by accident, in the street, at night or morning, that the patient perceives suddenly a numbness in the bottom of his feet, heaviness and feebleness in the inferior members, or some difficulty in walking. Our patient, Kelch, described these very characteristically. On rising in the morning, he first began to perceive that his feet were "asleep," that it was necessary to stamp them upon the floor for sometime before he was able to walk with any degree of certainty. This symptom was at first only experienced in the morning, but it gradually increased so as to be felt at all times. He attributed the uncertainty of his steps entirely to the sensation of numbness.

The progress of the malady is not always gradual; sometimes the symptoms occur suddenly and by leaps, as it were. When the symptoms of the first period have not appeared, or should they be incomplete, the beginning of the second period may be still either insidious or well-marked. Numbness, ataxia, anæsthesia, dysuria, and the same ocular phenomena may appear at the same time or at short intervals.

Numbness.—The ataxic allude under this name, to an internal sensation of heaviness, seated in a part or whole of a member, as if it were asleep. The numbness is a phenomenon, connected with the general sensibility, but distinct from the sense of touch, of heat, of pain, and from the muscular sensibility. It is a perversion of the functions of the sensitive nerves, as is the pains or the tingling. Some patients employ the words *numbness* and *tingling* as convertible terms. *Kelch*, so employs them in describing his sensations. The tingling, however, he refers to his forearms and hands, especially the right, more than to the inferior extremities. It corresponds to the distribution of certain filaments of the radial nerves and is accompanied by uncertainty of touch and inability to use his hands properly. As remarked in the history of his case, he had been unable for sometime to use his knife and fork. This sensation, however, did not extend to the trunk as it often does.

Cutaneous Anæsthesia.—Anæsthesia is one of the habitual symptoms of the progressive locomotor ataxia. In 109 observations, this symptom, complete or incomplete, was present in 76 cases, very lightly in 15, and not at all in 18 cases. It was very manifest in our patient, Kelch. A diminution, perversion, or exaltation of sensibility is found in this malady in the skin and in the mucous surface accessible to our investigation; but we have here in view the cutaneous anæsthesia, chiefly.

M. Beau recognizes two modes of sensibility in the integument—one special, or sensorial, as the sense of touch; the other general, as the sense of pain. M. Landry distinguishes a third, the sensibility to temperature.

When the anæsthesia is complete there is no difficulty; but if one or two of the three kinds of sensibility, only, is involved and that lightly, it is important to apply the best and most delicate tests. An examination of the ataxic to be complete should be extended to the four members, to the trunk and face, and to many points in the various regions, which habitually, are very unequally and differently affected. The prick of a sharp pin, pinching with the nails and with an artery forceps, plucking the hairs, tickling, the successive application of one or more fingers, of cold or hot bodies, the electric shock, may all be applied to determine the extent of the anæsthesia. When it is difficult to determine the modification of sensibility, comparison should be made between two symmetrical points, as the plantar surface of the feet, the inner faces of the thighs, the forearms, etc. Incomplete anæsthesia, as the numbness, advances step by step, from the plantar surface of the foot toward the pelvis and trunk. Its intensity is stronger in the part primitively affected, and its superior limit is not well defined. For example, the plantar surface of the foot, is without sensibility to touch, to pain or to tickling, the calf of the leg is somewhat more sensitive, the thigh is slow in transmission of the impression only, and the belly is normal. In the superior extremities the anæsthesia is experienced in the fingers, especially the ring and little fingers, diffuses itself over the rest of the hand and forearm, rarely passing above the elbow; but it reappears in the neck, in the inferior half of the face, and extends to the tongue, to the internal face of the jaws and to the vail of the palate. In our patient *Kelch*, this partial anæsthesia was confined to the hands and forearms—the ring and little fingers being more affected than the rest of these parts. The two halves of the body are not necessarily symmetrically affected. Thus one arm, one side of the face, of the tongue, is affected, while the other side may not be.

The sensibility to pain is diminished or abolished in places, in regions or in the whole extent of a member. The part affected may be pricked and the blood drawn without evidence of pain, but it is indispensable that the point of the instrument be exceedingly fine, for an ordinary pin will arouse the sense of touch. Pinching with the nails, which is easily graduated, has not this inconvenience. The sense of touch is habitually altered. The patient is deceived as to the real cause of an impression and as to the point at which it is made. He does not dis-

tinguish the touch of one or more fingers, from that of some other object. He confounds a small roll of paper with a pencil. The consequences of tactile anæsthesia may be, thus, attributed to ataxia.

Trousseau and Duchenne have observed that the thermoscopic sensibility continues in the ataxic. It was hence supposed that this constituted a means of differential diagnosis between progressive locomotor ataxia and other affections of the cord, but subsequent investigations have shown that this view is erroneous; the thermoscopic sensibility may exist in various diseases of the cord.

Muscular Anæsthesia.—The sentiment of muscular activity, the muscular sense, the muscular conscience, are synonymous terms, clearly separable, however, from the muscular sensibility.

Several methods may be adopted to ascertain the state of the muscular sensibility. The electrical method is mathematically accurate. It is applied as follows: the skin having been dried with some absorbent powder to neutralize the superficial effects of the electricity, a current of variable intensity is passed through a muscle in the direction of its length. In the physiological state this current determines a contraction, but in addition thereto, a dull sensation if it is feeble, and a pain if it is strong. This sensation—as was the case in our patient Kelch—whatever may be the intensity of the current, is in the ataxic, either very feeble or null. We thus measure the electro-muscular sensibility as it is called. The second method is more easy of execution. This consists in ascertaining the degree of sensation produced by pinching with the pulps of the fingers a sufficient quantity of the skin; afterward of a portion of healthy muscle, the sterno-mastoid, or biceps for example. This pressure graduated at will, may be made to produce acute pain. The suspected muscles are afterward explored in the same fashion. It is rare that the responses of the patient, how little intelligent soever he may be, leave any room for doubt. The third method is more especially to determine the state of the muscular sense. It is based upon this: In the physiological state, without having recourse to our sense of vision, we are concious of the direction which the will imposes upon our members, and of the precise point at which a movement is arrested. The use of the eyes is necessary to the ataxic, to conduct his movements

It is also useful to ascertain the degree in which the patient possesses the faculty of appreciating the weight of bodies, or the knowledge of the amount of muscular force necessary to raise a given weight. With this view different weights are placed in his hands, and his appreciation of them noted.

Locomotor Ataxia.—The locomotor ataxia is described in general

terms by Topinard, as a disorder of the function which presides over progression, equilibration, and other voluntary muscular acts, not caused by paralysis, muscular atrophy, deformity of the skeleton, etc., and different from paralysis agitans, chorea, or general or partial convulsions tonic or clonic.

The phenomena of the locomotor ataxia are to be studied as they manifest themselves in the inferior extremities, in the superior extremities, in the face, etc. Those partaining to the inferior extremities are most interesting and important. In our patient Kelch, as indeed in the greatest number of cases, the uncertainty of locomotion was most manifest in the morning in dressing. Sometimes it is felt in the street, more often in the evening and in darkness. They experience pain in mounting the stairs, or in stepping up on to the pavement. They continue to attend to business, but find they grow more readily fatigued than was their wont. A strong emotion, a slippery pavement, or the exertion to avoid an approaching carriage causes them to trip and fall unless there be some ready support at hand. Already they appreciate the utility of vision, and say that the disorders of locomotion are exaggerated because their attention is withdrawn from their own movements. To give them assurance in walking, they must have the idea that some one is near to succor them. The fears which they display, come of the experience which they have acquired of the instability of their movements and insufficiency of their support, and do not prove pusillanimity. Some authors have imagined that the defects of control of the voluntary movements are only experienced by the patient in the erect position; but they are also observed when he is in the recumbent position. When the patient gets under the bed clothes he extends himself by a quick and rough movement. If he be desired to execute a given movement, the muscles become rigid, the foot is extended upon the leg, the leg upon the thigh, and the limb, all of one piece, launches out at a single bound, or by irregular jerks, in the desired direction. The motions of the pelvic-femoral muscles, are most visibly ataxic. Intelligent patients explain the rigid extension which they instinctively give to the foot and leg in executing a given movement, by saying that their muscles are more rebellious to the will in intermediate attitudes.

When the patient is required to assume the erect posture, and place his feet together, he is seen to oscillate from side to side, betrays a certain anxiety, stretches out his arms like a ballancer, in spite of himself, and finally falls.

In walking the ataxia assumes considerable proportions, in many cases; in others, however, it is almost inappreciable. To facilitate the

description, we distinguish arbitrarily, in respect to intensity, three degrees of ataxia.

In the first degree the patient raises his feet roughly, one after the other; they describe the arc of a circle with the convexity turned out, without dragging upon the floor; the toes elevate themselves a little in advancing, and the heels turn in toward each other, striking the ground with force. He walks without decision, and can not place his feet where he desires.

In the second degree, the walking has been compared to that of a man inebriated, or to that of a landsman on a sailing packet, or to that of a rope dancer. An examination of the muscles discloses the fact that the flexors and extensors are hard and stiff. The triceps forms a considerable protuberance, which gives to the limb a special and forced configuration. The walk is carried on by the aid of the pelvic-femoral muscles. The feet are raised quickly, are jerked forward, the toes being elevated.

In the third degree of ataxia the patient is unable to step without holding on to a chair, to the bed post, or without being supported by two attendants. The sole of his foot seems in perpetual search of a point of support which continually escapes. Sometimes one of the feet turns into such a position that its outer side rests on the ground, and its dorsum appears in advance; or one leg, describing a great circle, becomes locked behind the external malleolus of the opposite limb.

It is difficult to describe the precautions and the stratagems which are used by those patients who have retained the use of their arms, in getting out of bed on to a chair and in pushing up to the stove. Their attention is applied to avoid the most feeble voluntary movements, they dare not abandon their caution for an instant, knowing that the first effort of muscular contraction will be the signal of an agitation which all their efforts can not repress.

In the superior extremities the ataxic disorders are less characteristic, and, also, less frequent. In *Kelch* they were marked and came on early. In 118 cases collected by Topinard, the ataxic disorders extended to the upper extremities in 46 cases, and in one only, were limited exclusively to these members. There are some differences, also, in the phenomena observed, due to certain differences in the physiological properties of the lumbar and cervical enlargements of the cord.

To discover the troubles of co-ordination of the superior extremities, many methods are employed. They were disclosed in *Kelch* by an inability to use his knife and fork. The patient may be desired to make the sign of the cross, to carry his index finger to the extremity of his

nose, to pick up some small object, etc. His movements may be examined when he eats, dresses, sticks a pin, ties a cravat, etc. These ataxic disorders appear in the upper extremity usually after they attack the lower; sometimes in the two extremities simultaneously. The first indication of them is seen in the awkwardness of the patient in performing the various acts above mentioned. He does not touch directly with the index finger, the point of his face which is designated to him. In all those acts in which the sense of touch is employed, and in picking up small objects, the ataxia becomes plainly visible.

In the third period, all of the symptoms of progressive locomotor ataxia have become general; the signs of the first and second period are united, much more intense and occupy a more extended surface. The movements are frightfully irregular and disordered in the erect position; prehension is impossible; cutaneous anæsthesia extends to impressions of heat and cold; the muscular anæsthesia is absolute; the muscular sense is abolished; there is incontinence of urine and fæces, and lastly, a true paralysis supervenes.

State of the Muscular force.—The ataxics are in general vigorous subjects, well nourished, whose muscles are hard and prominent, and limbs are voluminous. A small number, on the contrary, are irritable characters, hypochondriacs and feeble.

Before using the dynamometer to ascertain the state of the muscular force, the physician must study the constitution, temperament, idiosyncrasies of the subject, the lowering of his vital forces by mental depression, insomnia, bad hygiene, by the sojourn in hospital, etc. A simpler method than the use of the dynamometer, is that employed by Topinard, as also by Duchenne. For the muscles of the arm, shoulder, thigh, leg or foot, they should be placed in the attitude to insure their maximum contraction, and the patient should be directed to maintain, with all his force, this position; the degree of force necessary to overcome this resistance, will be a measure of the muscular force. It is sometimes necessary in the same subject to compare sound with affected muscles, or one side of the body with the other.

The fundamental condition of the ataxic paraplegia of Todd, and the progressive ataxia of Duchenne is the integrity of the muscular force. This proposition of Duchenne, is a little too absolute. Ordinarily the ataxia exists with persistence of a sufficient degree of muscular force to preclude the idea of paralysis; but, also, in a great number of cases there is a progressive feebleness, more apparent in the regions where the evidence of the other affections is most prominent, due to the influence of the cord upon nutrition and to an alteration of the motor parts

of this organ. Finally incomplete, and sometimes, complete paralysis is a remote termination of the malady. The anatomy and physiological pathology of this disease agree perfectly with this view. Considered with reference to the periods, it is certain that integrity of the muscular force is a character more constant in the second, although its diminution is an indication of aggravation of the lesion in the cord.

Course and Duration.—The course of the malady has been sufficiently indicated in the preceding pages. The duration of the first period only, can be approximatively fixed. We have before remarked that it oscillates between some months and twenty years; it averages from four to five years. From the origin of the disease, indicated with more or less clearness by the pains, the ocular troubles and the ataxia, to the second period—in 4 cases the duration is spoken of as months; in 6 cases as one year; in 27, as from two to three years; in 47, as from four to seven years; in 17, as from eight to fourteen years; in 18, as from fifteen to thirty years. The average is seven years. We have seen that in our patient Kelch, the duration of the first period, was somewhat less than one year—taking as the point of departure from the state of health, the sudden occurrence of spermatorrhœa. If, however, we assume that the ocular troubles were the first symptoms in his case, six months will include the whole course of the malady, so far as it had developed itself. There can be no question that the sexual disorder was the first symptom. It will be perceived that the phenomena observed in his upper extremities, were those of the first period, and not of the second. We have consequently no means of determining the duration of the first period if its evolution had not been interfered with it. As Topinard's statistics show, somewhat more than four per cent. had a duration less than one year.

Complications.—These are divisible into two classes: those pertaining to the cerebro-spinal axis; those of other parts of the body. Among the first are spinal meningitis, congestion and softening of the cord and of the brain, general paralysis, and progressive muscular atrophy.

Cephalalgia, dizziness and intellectual disorders which sometimes accompany the ocular derangements, must be viewed as complications. Some patients complain of loss of memory, and forgetfulness of certain words, certain events, etc. Others present various hallucinations. Mania or dementia, is found at an advanced period—the termination of the second or in the third period.

The complications of the second class are either acute or chronic. Of the acute affections, cystitis, enteritis and erysipelas are the most frequent. Cystitis usually results from the prolonged retention of the

urine—the bladder expelling it but in part. The symptoms first observed, are those of sub-acute, afterward of chronic cystitis. The inflammation extends along the ureter to the kidneys, and finally gives rise to urinous fever and putrid infection. The occurrence of enteritis, Topinard is disposed to ascribe to the prolonged use of the nitrate of silver, the remedy so much vaunted by Wunderlich in the treatment of the progressive locomotor ataxia. Of the chronic affections, phthisis pulmonalis is most common. In 44 deaths, 13 were attributed to this cause.

Forms.—The *common form* presents two distinct periods—the first and the second—or begins by locomotor ataxia, and has for characteristic the appearance, sooner or later, of functional disorders of the cranial nerves, of which the ocular troubles are the most important. In the *paraplegic form*, the affection is limited to the inferior members. To distinguish this from an ordinary myelitis, it is necessary to pay great attention to the progress of the case, to the physiognomy of the symptoms and to the characters of the ataxia.

In the *cerebral form*, cerebral phenomena are added to the symptoms of the common form, indicating morbid alterations in the brain corresponding to those affecting the cord and the periphery of the cranial nerves. Cephalalgia, changes of humor, loss of memory and delirium are the cerebral derangements characteristic of this form.

Our author strongly objects to the so-called *acute form* of this disease in which the total duration of it is limited to some months. The age of the subject (referring to one of these cases reported by M. Bourguignon), the rapid succession of the accidents, the total duration of four months and a half, the prompt and radical cure, are all in contradiction with the mass of our observations. He therefore rejects this form, but it seems to us, on insufficient grounds.

Pathological Anatomy.—The alterations characteristic of this disease have been carefully studied both by the naked eye and by the microscope. In the work of Topinard* very full and exact description of the morbid appearances is given. Trousseau† quotes with approbation from the essay of Dr. Axenfeld, *Des lesions atrophiques de la moelle épinière*. Having had no opportunity of studying these lesions for ourself, we propose nothing more than to condense from these authorities an account of the pathological anatomy sufficient to

---

* *Op. Cit.*, p. 310 to 387 inclusive.

† *Clinique Médicale de L'Hotel-Dieu de Paris. Tome deuxieme*, p. 534 *et seq.* Paris, 1865.

give some completeness to this essay, without running into prolixity of detail.

As a general rule the spinal meninges are free from disease; in a single case only in forty autopsies, according to Topinard, were they considerably affected. There is sometimes increased vascularity of the meninges, especially in the vicinage of the posterior columns.

The characteristic alterations are found in the posterior columns and in the posterior roots of the spinal nerves. These alterations consist in changes of form, volume, consistence, and color. The change in form and volume is due to atrophy of the columns affecting their antero-posterior diameter. There is usually a diminution of consistence, but in a few instances induration has been observed. The change in color is, according to Topinard, constant and almost pathognomonic. The columns become semi-transparent, of a gray color, variegated here and there of an amber yellow, which may become rose, according to the quantity of blood, and contrast strongly with the normal whiteness of the adjacent parts. The alterations in the posterior roots proceed correspondingly with the changes in the posterior columns. In Topinard's forty autopsies these roots were altered more or less in all but four cases. It is exceptional for this degeneration to invade the antero-lateral columns; it is ordinarily limited to the posterior columns.

There are no constant and characteristic alterations of cerebrum and cerebellum. The cranial nerves, whose functional derangement is a prominent feature in the symptomatology of this disease, are found to be more or less affected. In twelve cases carefully observed, two only, according to Topinard, presented no evidences of disease. The optic is probably more constantly affected than any other cranial nerve. The changes observed in it consist of an increase in vascularity, diminution in size and firmness, softening and atrophy, gray degeneration, and semi-transparency. Sometimes the atrophy is so considerable, that the nerve becomes transformed into a fibrous cord.

The description of the microscopic appearances by Dr. Axenfeld, quoted by Trousseau, is the most succinct, consistent with clearness, that has fallen under our observation. According to this author, the white substance of the posterior columns becomes gray or yellowish, and the nerve tubes, pale and shrunken, are sometimes reduced to the mere envelop containing granular contents. But few of them preserve the cylinder axis. An increase of the conjunctive hyaline substance in which the nerve tubes are implanted, gives to the nerve substance a fibrillar appearance; and there may be found here, in greater or less abundance, amyloid corpuscles, recognizable by their reaction with the

tincture of iodine. In the posterior cornua of the gray substance there occur the same alterations, but to a less marked extent. The changes in the posterior roots do not differ from those of the corresponding column. The vessels also are seen to be considerably enlarged, their coats thickened and incrusted with fat granules. "To sum up," says Axenfeld, "all of these alterations clearly consist in an atrophy of the nervous elements."

Topinard (p. 335) also, after a very elaborate discussion of the microscopic appearances, sums up his conclusions in a few words: "The microscopic examinations have shown the essential lesion in the different parts of the nervous system to consist of a gray, semi-transparent, or as it is styled by Cruveilhier and Virchow, a gelatiniform degeneration with a progressive atrophy of the nervous elements, hypergenesis of the conjunctive substance, amyloid bodies, and molecular granules."

Are the above described alterations constant to all stages of this disease? Is the progressive locomotor ataxia, in any part of its course, a functional disorder? At what period do these lesions of the posterior columns and roots manifest themselves? These are questions of peculiar interest. It has been said that the case of Kelch was not one of progressive locomotor ataxia, because the symptoms were evolved in so short a period of time, and were so much mitigated by treatment. Fortunately, the data exist for a solution of two of the problems proposed for our consideration. Let us take the testimony of Trousseau upon the first two questions. This ablest of modern physicians, in his lecture on this subject (vol. ii, p. 545–6), places this disease among the *neuroses*. He bases this opinion upon clinical observation, upon the nature of the symptoms which characterize the nervous troubles, upon the apyretic march of the symptoms, and upon the mode of evolution, the variety, and especially the mobility of the symptoms. "When," says Trousseau, "I assert that the malady is not subordinate to the existence of material lesions, I refer only to those lesions appreciable by our means of investigation." He quotes in support of this opinion the remarkable case observed by M. Gubler, in which, although the patient had been long afflicted with progressive locomotor ataxia, after death by variola, there was found *absolutely nothing* in the encephalon, in the medulla oblongata, and in the posterior columns and roots of the spinal cord. "The cord was, it is true, generally injected, but this may be explained by the paralysis supervening in the course of the variola." The optic nerves had undergone gelatiniform degeneration, and the *motores oculorum* were flattened, diminished in volume, but were not gray. In every other part of the nervous system the characteristic

alterations were entirely wanting. Topinard, who gives the same case (p. 353), concludes that the preceding observation (case of M. Gubler) tends to demonstrate, 1. That the symptom ataxia is not the immediate effect of the gray degeneration of the posterior columns; 2. *That it may exist six months before the degeneration of the posterior columns manifests itself*, that is to say, without anatomical alteration appreciable by our present means of investigation; 3. That in the evolution, not only of the degeneration, but of the disease considered in its totality, the visible alteration of the optic nerves is anterior to that of the cord. To obviate the difficulty in accounting for the phenomena observed in Gubler's case not dependent upon anatomical lesions, M. Topinard invents the words dynamical alteration (*altération dynamique*), which, we may be permitted to observe, express no understood pathological law or doctrine.

A variety of clinical facts support the conclusions of morbid anatomy. A certain number of cases of progressive locomotor ataxia, treated early, are either cured, arrested, or decidedly ameliorated. We have already alluded to the case of M. Bourguignon (Topinard, p. 298), in which there occurred complete recovery, the case having progressed through its several stages in a period of six months. Topinard also gives a case (p. 305) admitted to the Hotel-Dieu in the service of Professor Trousseau, in which very decided amelioration took place after one year and nine months of treatment. Dr. Radcliffe, physician to Westminster Hospital, in a recent lecture* on progressive locomotor ataxia, speaks of "two cases which justify a much more hopeful opinion" than is usually prognosticated. D. Meryon† gives a case for which he advised the twentieth of a grain of strychnia to be taken night and morning, and the employment of very weak cutaneous faradisation, in which, after five months' perseverance, "there is decided improvement." Further, the great apostle of this disease, Duchenne, affirms that he has cured by faradisation cases in his first stage. We may safely add to this array of testimony our own observation. What is the present state (May 1) of our patient, Kelch? The amelioration in all of his symptoms, described in the first part of this paper, continues, but he has not entirely recovered. A certain degree of ataxia remains, and is especially manifest when he rises from his chair and attempts to walk, or when he turns without using his eyes to direct his

* *London Lancet*, January, 1866.

† Practical and Pathological Researches on the various forms of Paralysis. London, 1864.

feet. Thus far we can only affirm, with regard to his case, that it is one of "decided improvement."

A careful study of the clinical facts and *post mortem* observations authorize the conclusion that in the beginning the progressive locomotor ataxia is a functional disorder, and that a certain number of cases, if not curable, are capable of very great improvement. The boundary between functional disorder and organic change is unfortunately not at all clearly marked in our clinical observations, and not well defined even by our present means of investigations in morbid anatomy. When does the degeneration commence in the posterior columns? The symptoms do not enable us to decide this question. Topinard, as we have seen, asserts that the ataxia may exist six months before any but a "dynamic alteration" occurs. The division of Duchenne into the three periods based upon the symptomatology is, therefore, an arbitrary arrangement.

Division into Periods based upon the Pathological Conditions.—In this view, the *first period* is one of functional trouble merely: the *second period* one of organic change. The first period is characterized by the mobility and intermittence of all the symptoms, their spontaneous disappearance, or the decided amelioration of them by treatment; the second by the persistence and progressive increase in the gravity of all the symptoms, especially the amaurosis and the difficulty in the co-ordination of voluntary movements. All of the phenomena in our patient, Kelch, would, therefore, be referred to the First Period, except the ataxia, which, persisting, probably inaugurates the Second Period.

Differential Diagnosis.—There can be little difficulty in the diagnosis of a pronounced case of progressive locomotor ataxia. During Duchenne's first period—the stage of functional disorder—it is not always possible to interpret the pains, the ocular troubles, and the anaphrodisia correctly. It is very important to recognize early the import of these symptoms, which may be done by attention to the order in which they occur, to their temporary character, and to their unaccountable disappearance. As the disease progresses the difficulties in diagnosis diminish. The particular maladies with which it is most liable to be confounded are myelitis, softening of the brain, and general paralysis. The pains, the muscular anæsthesia, the persistence of the thermoscopic sensibility, the urinary and genital troubles, the ataxia, and the persistence of the muscular force, clearly distinguish this disease from those above mentioned, in which these symptoms are wanting. Some excellent observations on the differential diagnosis

have been made by Althaus, Hughlings Jackson, Radcliffe, and others in the *London Lancet* during the past year. All of these observers allude especially to the ataxia and to the persistence of the muscular force as diagnostic.

ETIOLOGY.—Topinard (p. 371), after a careful analysis of one hundred and nineteen cases, sums up his conclusions as to the causes of this disease in the following terms: "The progressive locomotor ataxia is a disease of mature age, more frequent in the male sex, in those who are addicted to excesses of all kinds, and who are exposed to humidity and to great fatigue. Among the temperaments, the nervous is the only one which certainly disposes to a particular form of the malady. Among the diatheses, the rheumatismal is the only one the influence of which is incontestible." Trousseau (vol. ii, p. 514) ascribes considerable importance to hereditary tendency. It will be found in many cases, he says, that the direct or collateral ancestors have been affected with other forms of nervous diseases.

The relation of sexual excesses to the production of this disease is an interesting question. We have already presented the opinions of Topinard on this point. The views of many of the German pathologists of the causes producing *tabes dorsalis* have descended from Hippocrates. "Tabes dorsalis," says Hippocrates,* "arises from the medulla spinalis, and happens chiefly to those who are libidinous or lately married," etc. Celsus† alludes to similar effects caused by *nimia profusio seminis*. Van Swieten‡ repeats the observations of Hippocrates. Lallemand,§ in his description of the "dorsal consumption," speaks of troubles of vision, amblyopia, diplopia, anæsthesia, and difficulties of locomotion, etc., resulting from spermatorrhœa. "It is impossible," says Dr. Meryon (*op. cit.*, p. 197), "to ignore the resemblance between this disease and that which is known as tabes dorsalis. Both may be occasioned by onanism or by excessive venery; both give evidence to the existence of irritation in the spinal cord; in both enuresis and spermatorrhœa frequently occur," etc. This view of the influence of sexual disorder can hardly be admitted. The venereal excesses and the spermatorrhœa must be regarded as symptoms, chiefly, of the mischief in progress in the spinal cord. Trousseau (*loc. cit.*) gives some striking facts in proof of this; cases in which the singular faculty existed of

* De Morbis, lib. ii, cap. 19. I have already remarked that Dr. Adams assigns this work to the Cnidian school.

† De Medicina, lib. iv, p. 187. Edit. Ed. Milligan. Edinburgh, 1831.

‡ Commentaries, vol. v. p. 85.

§ Dictionnaire de Médecine. Art. Spermatorrhée, par M. Raige-Delorme.

being able to repeat the sexual act frequently in a short space of time; "a deviation from the physiological state." One of these patients admitted to having intercourse six or seven times in twenty-four hours. The first symptom in our patient, Kelch, was a remarkable increase in the activity of his sexual desires; this, however, was quickly followed by profuse spermatorrhœa.

Was there any thing in the occupation or external conditions of our patient, Kelch, to account for the production of his symptoms? In his business, as a gilder, gold foil is the substance chiefly employed, and a small quantity of white lead in the preparation of the frames. The symptoms have been ascribed, by some persons unacquainted with the essential phenomena of the progressive locomotor ataxia, to lead poisoning. We might adduce, in answer to this doubt, two cases quoted by Topinard, in the first of which (ob. xxxiii) the symptoms were clearly those of lead poisoning (*intoxication saturnine*), but in the second, a painter (ob. cxcviii), all of the phenomena were those of the progressive locomotor ataxia. Kelch had never suffered from colic, paralysis, nor did he present any of the evidences of mineral poisoning, except the symptoms of the disease under which he labored, which may or may not be attributed to his occupation; most probably not. There is no evidence of hereditary tendency in his case.

Treatment.—The treatment of the progressive locomotor ataxia is a most unsatisfactory subject. The term progressive is unfortunately too often most fitting; for no matter what therapeutical expedients we may adopt, the disease continues on its course. Much depends, however, as we have already seen, upon the period of the disease in which we find our patient. The very unfavorable prognosis ordinarily expressed, heretofore, must now be somewhat modified. The cases which we have adduced show that appropriate treatment may accomplish much, if employed during the stage of functional derangement.

Duchenne relies upon the iodide of potassium and faradisation. We have quoted two cases from the *Medical Times and Gazette*, in which the use of the iodide was followed by improvement. Dr. Hughlings Jackson says that this drug was of considerable benefit in one case under his charge, and without service in others. Our own case is a striking example of its value.

The nitrate of silver, proposed by Wunderlich, has been much employed with apparent advantage in some cases, but in many more it has failed utterly. The propriety of prolonged administration of this remedy is very questionable; at most the benefit is very doubtful, and

its ill results are certain. Topinard attributes to this remedy the enteritis which has been observed in some cases.

Ergot and belladonna in combination proved very successful in the hands of Dr. Charles Taylor. Trousseau thinks favorably of the latter. Arsenic, oil of turpentine, and strychnia have also been used without any very decisive results.

The favorite remedies with the French are sulphur baths, douches, and electricity—cutaneous faradisation chiefly.

An improved hygiene is very important. The subjects of this malady are principally those who have been exposed, by reason of poverty and unhealthy occupations, to the common causes of disease. A more abundant diet, a purer atmosphere, warmer clothing, and cleanliness have in many instances apparently arrested the progress of the disease. These hygienic means should not be neglected in any case. Without their aid remedial agents will probably prove useless. Certainly the facts which we have presented in the course of this paper give us encouragement to persevere in the treatment, for, if not curable, we have every reason to hope for an arrest of the morbid action, an improvement in the symptoms, or a decided amelioration in all of the attendant phenomena.